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# Bilateral Primary Non - Hodgkin's Ovarian Lymphoma in a Young Female - A Case Report

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**Abstract:** ***Primary ovarian lymphoma is a rare entity. Here, we report a rare case of bilateral primary ovarian lymphoma in a 28 - year - old female, who presented with left - sided lower abdominal pain. MRI of the pelvis revealed bilateral ovarian masses. The patient underwent staging laparotomy with a total abdominal hysterectomy and bilateral salpingo - oophorectomy. The diagnosis of Non - Hodgkin's Lymphoma of bilateral ovaries was made on histopathological findings and immunohistochemistry. The final diagnosis of bilateral primary Non - Hodgkin's ovarian lymphoma was based on diagnostic criteria by Fox et al. The rare incidence of this lesion at young age with unusual imaging findings warrants its mention.***

**Keywords:** Non - Hodgkin's Lymphoma, ovarian tumor, malignant lymphoma

## 1. Introduction

Although Non - Hodgkins lymphoma frequently involves the ovaries at autopsy, they rarely involve the ovaries at initial presentation [1]. The occurrence of lymphomas involving the ovaries primarily is debatable because lymphocytic aggregates are never found in the normal ovary, although they may occur in association with the ovarian lesions of chronic pelvic inflammatory disease. Lymphocytic foci in pre - existing ovarian teratomas represent another possible source for the development of primary malignant lymphomas [2]. Here, we report a rare case of a young female with primary bilateral ovarian lymphoma.

## 2. Case Report

A 28 - year - old unmarried, nulligravida, nulliparous female presented with a history of pain on the left side of the lower abdomen of 1 - month duration which was insidious in onset, mild to moderate in intensity, dull aching in character, non - progressive & non - radiating in nature. There was no history of any associated systemic complaints. She had regular menstrual cycles with normal amount and duration of flow. She did not have any significant co - morbidities.

On per abdomen examination, she had a large abdominopelvic mass which was firm to hard in consistency. Per vaginum examination revealed bilateral adnexal masses.

An abdominopelvic ultrasound revealed multiple masses filled in the pelvis extending to the upper abdomen with mild hydronephrosis of the right kidney and mild ascites. A contrast - enhanced MRI of the pelvis was done which revealed two heterogeneous masses possibly arising from bilateral ovaries, iso to hyperintense on T1 weighted and hyperintense on T2 weighted images; left - sided mass= 12x13x17cm abutting uterine fundus and right - sided mass= 13x1x16cm. On the basis of imaging studies, differential diagnoses made were; 1. Germ cell tumor (dysgerminoma, mixed malignant germ cell tumor) and 2. Fibroma

Baseline serum markers (beta HCG and AFP) were normal whereas LDH was 287 IU/L.

A provisional diagnosis of bilateral ovarian tumors was made.

The patient underwent a staging laparotomy with total abdominal hysterectomy with bilateral salpingo - oophorectomy and a midline incision. Intraoperatively, there was a solid, irregular mass (8x10cm) over the right - side ovary, another similar mass (10x12cm) over the left side ovary, and two masses in the left broad ligament (5x3cm) invading the ureter. No normal ovarian tissue could be seen. The left ureter was slightly dilated with tumor infiltration in the wall. Therefore, per op diagnosis of bilateral ovarian masses with secondaries in broad ligament and ureteric infiltration was made.

Frozen section from right ovarian mass and omentum revealed malignant round cell tumor with differential diagnoses of germ cell tumor favoringdysgerminoma or Non - Hodgkins lymphoma.

On gross examination, the right adnexal mass measured 17x12x7cm and was solid, and fleshy with focal areas of hemorrhage; the left adnexal mass measured 15x12x8cm and was yellow, lobulated with hemorrhagic specks.

Microscopic examination of bilateral adnexal masses showed diffuse and solid proliferation of atypical round cells with round to oval vesicular nuclei with few showing prominent nucleoli with a scant amount of cytoplasm. Mitotic figures (2 - 3/HPF) imparting a starry sky

appearance were seen. Areas with capsular invasion were also seen. No lymphovascular invasion was identified. Bilateral fallopian tubes and cervix were unremarkable while the uterus showed serosal and myometrial infiltration by atypical cells. Myometrium also revealed a spindle cell neoplasm. Omental tissue was also infiltrated by similar atypical cells.

Immunohistochemistry showed membranous positivity in tumor cells for CD19 and CD20.

The final diagnosis based on histopathological examination and immunohistochemistry was made as malignant small round blue cell tumor favoring Non - Hodgkins lymphoma of bilateral ovaries with capsular invasion with involvement of uterus and omental tissue with leiomyoma of the uterus.

The patient was further evaluated by whole - body PET - CT scan for staging which revealed left para - aortic and left common iliac lymph nodes with significant FDG uptake.

The patient was staged as stage IV according to Lugano classification and is planned for systemic chemotherapy with an R - CHOP regimen comprising rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone.

## 3. Discussion

The gynecologic tract is an uncommon site for non - Hodgkin's lymphoma (NHL). However, when involved by NHL, the ovary is one of the common anatomic sites [3]. Dimopoulos et al reported that primary ovarian Non - Hodgkins lymphoma constitutes only about 0.5% and 1.5% of all Non - Hodgkins lymphoma and ovarian neoplasm cases respectively [1]. Fewer than 1% of patients with malignant lymphoma present with ovarian enlargement. Most lymphoid neoplasms of the ovary are Non - Hodgkins type. Malignant lymphomas involving ovaries are usually B - cell neoplasms [4]. DLBCL is the most common histologic subtype followed by Burkitt'slymphoma [3]

Ovarian lymphoma is mostly seen in women in the 4th decade of life but can occur at any age. Vang et al in their study observed patients' age ranged from 29 to 62 years with a mean age of 47 years [3]. Our patient presented at an age of 28 years which is an unusual presentation. Vang et al. reported that pelvic or abdominal complaints were the most common symptoms however in one - third of cases it was an incidental finding [3].

The presence of normal lymphoid tissue in the ovaries is controversial. While Nelson et al. stated that lymphoid tissue is not present in the ovaries [2]; Woodruff et al on the contrary stated that lymphoid aggregates may be found in the hilus and medulla of normal ovaries [5]. Monterosso et al detected well - defined aggregates of lymphoid tissue in the normal ovary [4].

Malignant lymphoma may involve the ovary as (a) a primary neoplasm arising in the ovary or (b) secondarily, as the initial clinical manifestation of occult nodal disease, or as a manifestation of widely disseminated systemic lymphoma [4].

Bilaterality is infrequent but in our case, the uncommon bilateral ovarian disease is seen [3]. The propensity for bilateral ovarian and extensive intraperitoneal involvement without extra - diaphragmatic involvement can probably be explained by the emergence of ovarian lymphoma by the malignant transformation of normal ovarian lymphoid tissue, and probably reflects the underlying homing and trafficking patterns of these normal ovarian lymphocytes [1].

Fox et al. in 1976, proposed the following criteria for the diagnosis of POL: (i) At the time of diagnosis, the lymphoma is clinically confined to the ovary and a complete investigation fails to reveal evidence of lymphoma elsewhere. However, an ovarian lymphoma can still be considered primary if it has spread to immediately adjacent lymph nodes or if it has directly spread to infiltrate immediately adjacent structures. (ii) The peripheral blood and bone marrow should not contain any abnormal cells. (iii) If further lymphomatous lesions occur at sites remote from the ovary, then at least several months should have elapsed between the appearance of the ovarian and extra ovarian lesions [8]. Our case meets all the criteria proposed by Fox et al and is therefore diagnosed as a case of primary ovarian non - Hodgkins' lymphoma (considering para - aortic and left common iliac lymph nodes as adjacent lymph nodes and involvement of uterus and omental tissue as direct spread to adjacent structures).

Ferrazzi et al observed ovarian lymphomas were frequently bilateral and homogeneous, and the lesion size always exceeded 5 cm in diameter on radiological imaging. CT scans showed hypodense distinct lesions with mild contrast enhancement. Ultrasonography showed nonspecific but homogenous and hypoechoic patterns. Magnetic resonance imaging analysis showed homogeneous masses that have moderately hypointense T1 - weighted images and slightly hyperintense T2 - weighted images [6]. On the contrary, MRI analysis in our patient revealed heterogeneous and hyperintense bilateral ovarian masses on T1 and T2 weighted images which is an unusual finding and could lead to misdiagnosis as other ovarian neoplasms.

The histological appearances of lymphoma in the ovary are generally similar to those seen in the extra - ovarian sites. In the ovary, however, there is a great tendency for the tumor cells to grow in cords and nests, appearing to cling to the reticulin, forming pseudoacini [9].

Most patients with ovarian lymphomas are treated with surgery followed by chemotherapy, whereas treatment by radiotherapy is less common. A CHOP regimen (cyclophosphamide, adriamycin, vincristine, and prednisone) is often administered because CHOP is the most standard regimen for the treatment of non - Hodgkin lymphoma [7]. Rituximab plays an important role in the management of CD20 - positive Non - Hodgkin's Lymphomas [10].

## 4. Conclusion

Primary ovarian lymphoma is a rare entity. It can be easily misdiagnosed as epithelial ovarian malignancy owing to similar presentation. Differentiation from other ovarian neoplasms is a must as the management and prognosis are significantly different. Keeping primary ovarian lymphoma as a possible differential diagnosis while evaluating a patient with ovarian masses can prevent the patient from undergoing unnecessary radical surgery. Chemotherapy should be the main treatment option. Early diagnosis and prompt treatment can further lead to improved survival rates and quality of life of the patient.

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